

SLE – Systemic Lupus Erythematosus

Definition

-Lupus is a chronic multisystem inflammatory disease characterized by remissions, and acute or chronic relapses.

Epidemiology

- Higher incidence in females, African American, age 20s-30s
- Prevalence approximately 40-50 cases out of 100,000

Common Symptoms – Remember our mnemonics? I'M DAMN SHARP or SOAP BRAIN MD if it helps

-Constitutional –fatigue (74-100%), fever (40-80%), weight loss, arthritis/arthralgia (83-95%), skin (rash, alopecia, Raynaud's), renal, GI, pulmonary, cardiac, lymphadenopathy, splenomegaly, hepatomegaly, CNS (seizures, psychosis, mental status changes)

Diagnosis – Only get ANA titer if patient presents w/manifestations involving >2 organs

Positive ANA not diagnostic, but negative test argues strongly against SLE

ACR Criteria – 4 of 11	SLICC Criteria – 4 of 17 (1 clinical + 1 immunological, OR biopsy proven lupus nephritis)
Malar rash	Acute cutaneous lupus
Photosensitivity	Chronic cutaneous lupus
Discoid rash	Non-scarring alopecia
Oral ulcers	Oral or nasal ulcers
Arthritis – 2 or more joints	Joint disease
Serositis- pleuritis or pericarditis	Serositis
Renal disorder – proteinuria or casts	Renal involvement
Neurologic disorder	Neurologic involvement
Hematologic disorder	Hemolytic anemia
ANA titer \geq 1:40	Leukopenia or lymphopenia
Immunologic Disorders	Thrombocytopenia
	ANA
	Anti-dsDNA
	Anti-Sm
	Antiphospholipid
	Low complement
	Direct Coombs' test

Testing - Base frequency of testing on severity of disease

- Baseline testing and help with diagnosis include CBC, CMP, CK, ESR, U/A, urine protein:creatinine ratio, ANA, antiphospholipid antibodies, anti-dsDNA, anti-Sm
- During flares, low complement levels, and if renal involvement check frequent U/A and NKBC

Treatment

- Sun protection, diet, exercise, smoking cessation
- Treat specific organ involvement: Topical therapies for cutaneous involvement, NSAIDs for constitutional sx
- Antimalarials for skin and msk (i.e. hydroxychloroquine)
- Systemic glucocorticoids alone or in combination
- Immunosuppression (i.e. methotrexate, cyclophosphamide)
- In resistant disease, time to consult the specialist

References

- Gill, et al. Diagnosis of Systemic Lupus Erythematosus. *AAFP*. 2003;68,11: 2179-2186
- Schur et al. Multiple articles on Up To Date edited in January 2013.